



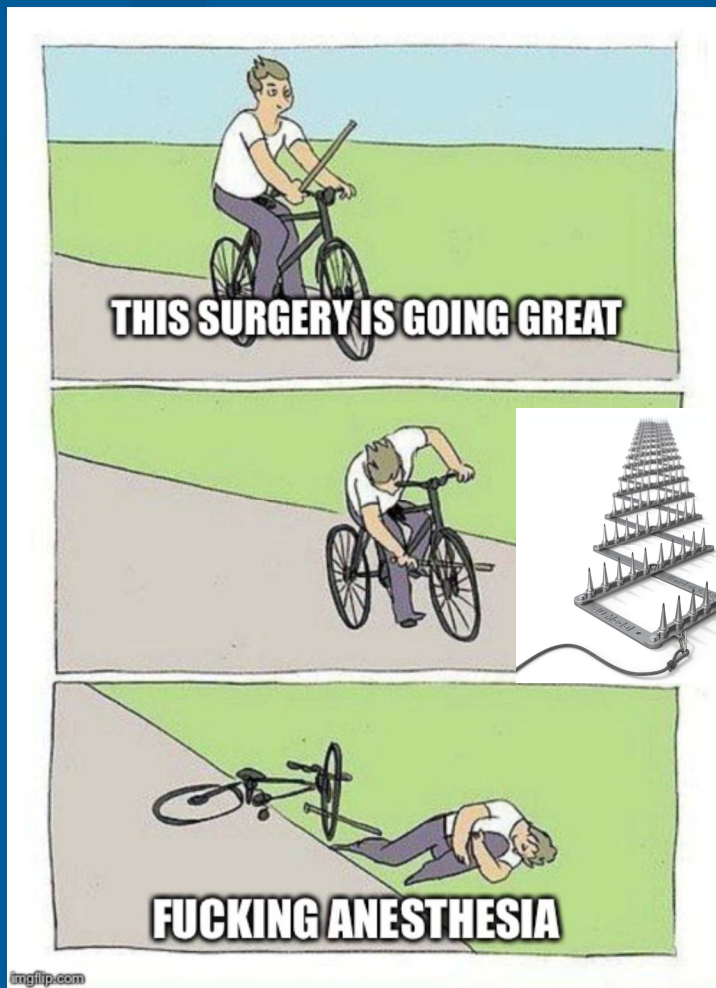
Anesthesia and Early Onset Scoliosis Surgery: Minimizing Risk In Syndromic Patients?

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Disclosures

- a. Grants/Research Support
- b. Consultant
- c. Stock/Shareholder
- d. Speakers' Bureau
- e. Editorial/Governing Board
- f. Other Financial Support

Lydia E. Andras MD - Eli Lilly (c)



Syndromic Patients

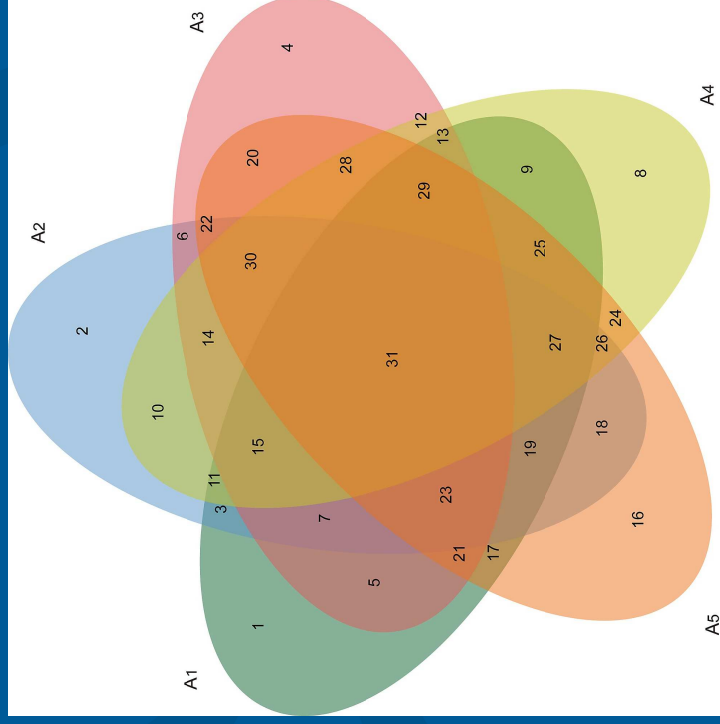
Anesthetic Safety

- Serious adverse events 1.4 per 1000 anesthetics
 - Airway/Respiratory #1
 - Cardiac arrest #2
 - Medication error/equipment #3
- 30,000 pediatric anesthesia MRI sedations from Pediatric Sedation Research Consortium, zero deaths, 5.3% brief oxygen desaturation
- For syndromic EOS patients these numbers are likely higher: ASA 3 and 4, long surgery, hemodynamic shifts, multiple comorbidities

BE PREPARED

IT'S NOT
JUST FOR SCOUTS





Anesthetic Safety in EOS Surgery for Syndromic Patients

- Airway
- Cardiac
- Respiratory
- Drug reaction
- Neurotoxicity
- Positioning

I am not anxious.
I am just extremely well
educated about all
the things that
can go
catastrophically
wrong.



someecards
user card

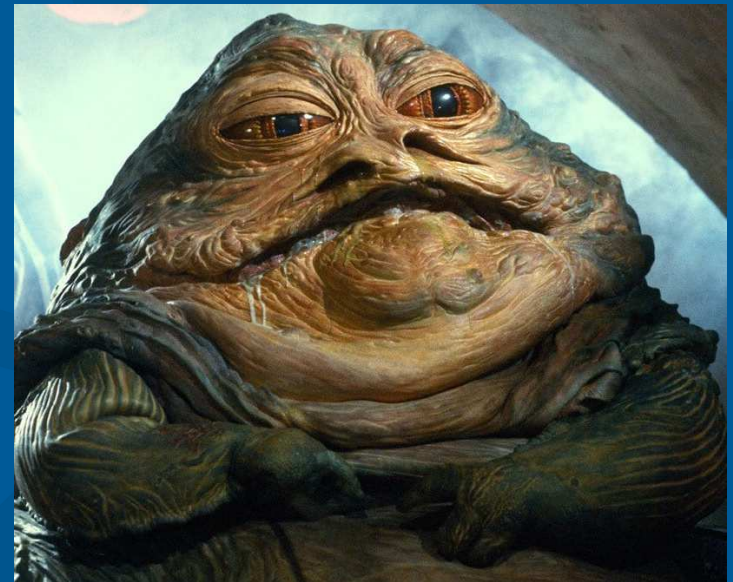
Airway

- Ventilating
- Intubating
- Avoiding Aspiration
- Extubating



Airway

- Screening Syndromic Patients' Airways
 - Limited neck mobility/Thick neck
 - Limited mouth opening or small mouth
 - Large tongue
 - Short chin
 - History of previous difficult intubation/ventilation
 - Mallampati vs COPUR (Chin, Opening, Previous difficult intubation/OSA, Uvula visualization, Range of opening)
 - Known Offenders:
Mucopolysaccharidoses, Trisomy 21, Apert, Klippel-Feil, etc.



Airway

- Experienced providers
- Airway Adjuvants
 - Video laryngoscopy, Fiberoptic, Intubating LMAs
- ENT available
- Schedule Wisely
 - First case, weekday, extra time



Airway

- Aspiration
 - Increased risk in patients with decreased esophageal tone, delayed gastric emptying
 - NPO violations- DELAY!
 - Clears- **2** hours
 - Breast milk- **4** hours
 - Formula/light meal- **6** hours
 - Full meal- **8** hours
 - Commonly seen with: Rett, Angelman, obesity (Prader Willi, Trisomy 21)



Airway

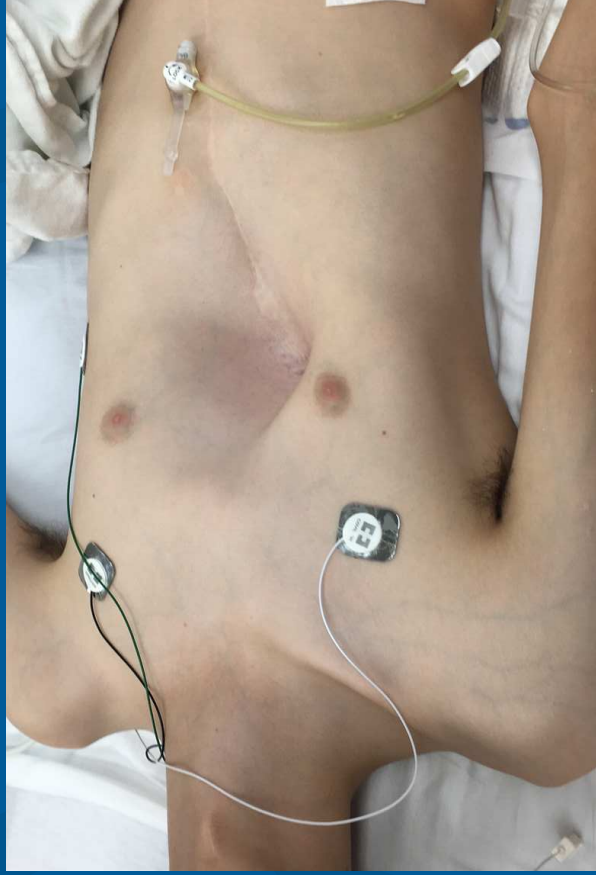
- Extubating
 - For: decreased risk of pulmonary infection, decreased likelihood of tracheal stenosis, sedation not required, don't HAVE to go to ICU
 - Against: Reintubation (harder), risk of acute respiratory events, not able to support ventilatory status in high risk time



**KEEP
CALM
AND
EXTUBATE
MAYBE**

Cardiac

- Patient's intrinsic risk-Are they optimized?
 - Congenital heart disease: VACTERL, Trisomy 21, Marfan, Noonan, Ehlers Danlos
 - Pectus: Marfan, Ehlers Danlos, Noonan, OI
 - Conduction abnormalities: Rett(QT), Angelman and Trisomy 21 (bradycardia)
 - Myopathy: Muscular dystrophies
 - Pulmonary Hypertension/Right heart failure: Depending on curve and comorbidities many!
 - Cardiovascular Disease: Prader Willi
 - SBE prophylaxis: Dependent on diagnosis but certain subgroups at increased risk





[Pediatric Cardiology](#)

October 2019, Volume 40, [Issue 7](#), pp 1468–1475 | [Cite as](#)

Adverse Perioperative Events in Children with Complex Congenital Heart Disease Undergoing Operative Scoliosis Repair in the Contemporary Era

Authors

[Authors and affiliations](#)

Robert Przybylski , Daniel J. Hedequist, Viviane G. Nasr, Mary Ellen McCann, Robert M. Brustowicz, John B. Emans,

Audrey C. Marshall, David W. Brown

- Single Institution- 78 patients
- ~20 % complication rate
- No Deaths
- Large Cobb angle and cyanotic single ventricles highest risk

Cardiac

- Surgical Risk- minimize what you can:
 - Controlled Hypotension BUT not in cardiomyopathy, single ventricle, unstable patients
 - Antifibrinolytic (TXA)
 - Blood available (may take longer, may need more than you think you need)
 - Rapid Transfusion available
 - Staging



Respiratory

- Restrictive lung disease: Many!!
- Pneumothoraces: connective tissue disorders
- Recurrent Pulmonary Infections: Trisomy 21
- Obstructive Sleep Apnea: MANY! Trisomy 21, Prader-Willi (can be worsened with GH), Hunter/Hurler



Respiratory

Sleep Apnea:

Severity of OSA	Adult AHI	Pediatric AHI
None/normal	0-5	0
Mild OSA	5-20	1-5
Moderate OSA	20-40	5-10
Severe OSA	>40	>10

J Intensive Care Med. 2017 Mar;32(3):204-211. doi: 10.1177/0885066616661152. Epub 2016 Aug 20.

Analysis of Unplanned Intensive Care Unit Admissions in Postoperative Pediatric Patients.

Landry EK¹, Gabriel RA², Beutler S¹, Dutton RP³, Urman RD¹.

- 324,818 cases analyzed, 211 unplanned ICU admissions
- Risk Factors: age less than 1 year, ASA 3 or 4, longer surgery, surgery with general anesthesia

Drug Reactions

- Anesthetic Gasses/Succinylcholine
 - Malignant hyperthermia
 - King Denborough, Multiminicore, Central core, Nemaline Rod myopathy (certain variants), RYR1
 - Rhabdomyolysis/hyperkalemia
 - Muscular dystrophies



Drug Reactions



Malignant Hyperthermia Association of the United States

24-HOUR MH HOTLINE
800-644-9737
Outside NA: 001-209-417-3722
FOR EMERGENCIES ONLY

- www.mhaus.org
- First Cases (or be prepared for delay)
- Delayed emergence

Drug Reactions

- Anaphylaxis
 - 1:10,000...likely more
 - Antibiotics, Neuromuscular Blockers, Chlorhexidine, Latex
 - Hypotension, Bronchospasm, Urticaria, Angioedema
 - Secure airway...if not already intubated can make a tough job tougher

BJA

British Journal of Anaesthesia, 121 (1): 159–171 (2018)

doi: 10.1016/j.bja.2018.04.014

Advance Access Publication Date: 21 May 2018

Special Article

Anaesthesia, su
reactions: epide
perioperative a
Project (NAP6)

N. J. N. Harper^{1,2,3}, T.
S. Marinho⁹, H. Torev
W. Egner^{12,13}, H. Kem
S. Karanam¹⁸, K.-L. Ko



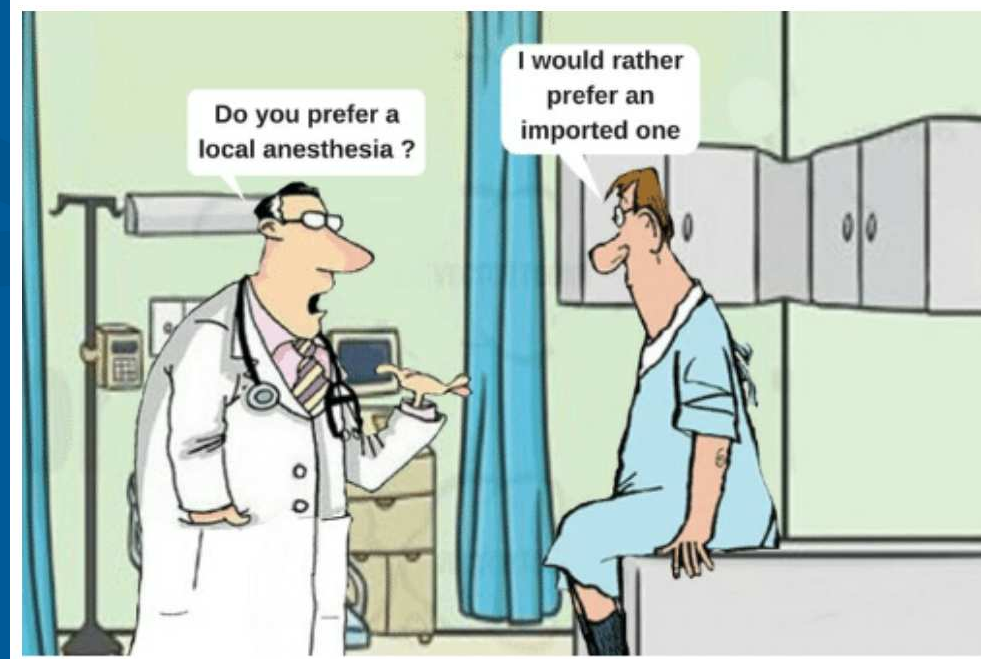
Sources: Goldsmith LA, Katz ST, Gilchrist BA, Paller AS, Jeffell DJ, Wolff K. Fitzpatrick's Dermatology

Allergies: amoxicillin->throat swelling, gold->skin turns green

Neurotoxicity



Neurodevelopmental outcome at 5 years of age after general anesthesia or awake regional anesthesia in infancy (GAS):



it,
rowley,
Szmuk,

by neuronal
association
s are prone
esia in early

Age

MPH,
ays, MD,
oun,



Spine Deformity

Volume 6, Issue 6, November–December 2018, Pages 781-786



Case Series

Cumulative Anesthesia Exposure in Patients Treated for Early-Onset Scoliosis

Fady J. Baky BS ^a, Todd A. Milbrandt MD, MS ^a, Randall Flick MD, MPH ^b, A. Noelle Larson MD ^a 



- Early presentation and treatment with casting more likely to have over 3 hours before 3 years of age

SPINE. 44(12):866-871, JUNE 15, 2019

DOI: 10.1097/BRS.0000000000002966, PMID: 30540716

Issn Print: 0362-2436

Publication Date: June 15, 2019



 Print

Limited Sequence MRIs for Early Onset Scoliosis Patients Detected 100% of Neural Axis Abnormalities While Reducing MRI Time by 68%

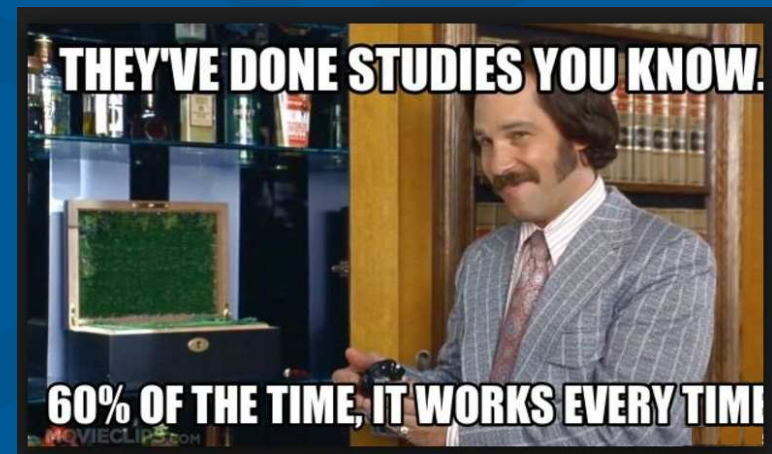
Rajan Murgai;Benita Tamrazi;Kenneth Illingworth;David Skaggs;Lindsay Andras;

- Less Anesthesia Time=Less Risk



Positioning

- Much be extra cautious:
 - Fractures- OI (BP cuff, wake up, bed rails)
 - Injuries due to hypermobility: Ehlers Danlos, Marfan
 - Pectus- chest rolls
 - Injuries due to instability: more to come in the next talk 😊





No pressure on the
central portion of
the chest

Consider
intraoperative
TEE

Positioning



- Communicate with the team
- Fix what you can, optimize what you can, and be prepared for what you can't



